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Music therapy in Huntington's disease

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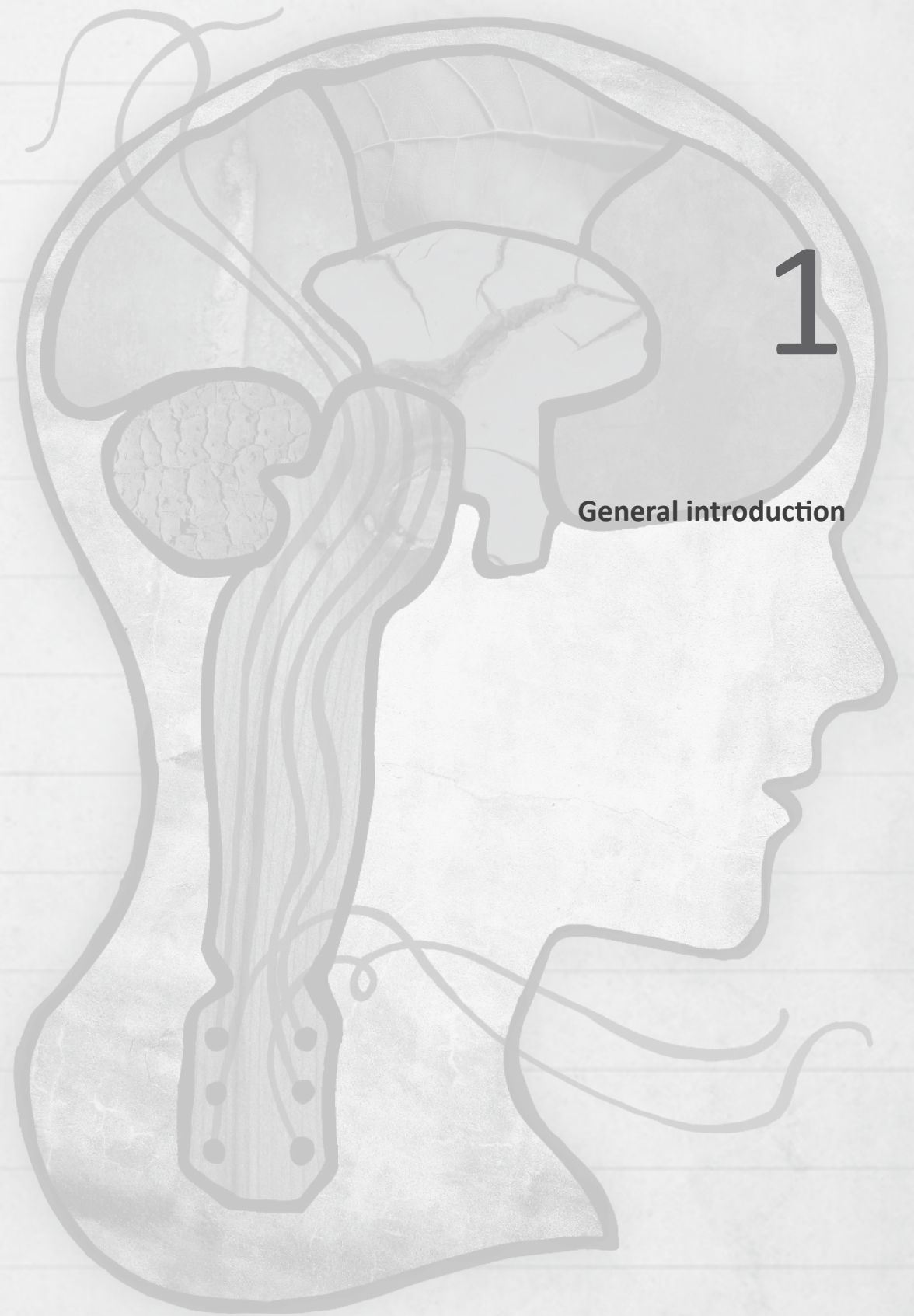


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General introduction

Huntington's disease

Huntington's disease (HD) is an inherited rare autosomal dominant disorder, caused by a mutation in the HTT-gen, causing progressive neural degeneration of the basal ganglia and gradual atrophy of frontal and temporal cortex [1]. The disease is characterized by movement disturbances, neuropsychiatric disturbances and cognitive deterioration leading to a complete dependency for daily life activities [2]. Clinically, a great variation in age at onset, duration of illness, and course of the disease is seen. The mean age of onset is between the age of 30 and 50 years, and once manifest, the patient's life expectancy is about 15-20 years [3]. Three clinical stages can be described: Stage I, in which patients develop initial symptoms but are still independent. Stage II, patients become more dependent as the disease progresses. Stage III is the end-of-life stage: patients become completely dependent for all daily life activities [2,3]. The prevalence of HD in Europe, North America and Australia is about 10 per 100.000 [4]. In the Netherlands there are approximately 1,700 persons with HD.

Communication problems in HD

The characteristics mentioned above often result in loss of expressive and communicative skills, especially in the advanced stage of the disease. Verbal communication is often affected, due to motor impairment of speech. In addition, as a result of the cognitive decline, word retrieval is often impaired, although the knowledge of vocabulary is retained. As the disease progresses, the language content can no longer be processed properly and adequate communication becomes challenging [1].

The gradual deterioration of the communication skills contributes to a sometimes rapidly increasing inability to participate in different life situations [5]. The communication problems may have a great impact on the quality of life of the patients and formal and informal carers.

Behavioral problems in HD

Behavioral problems are common among patients with HD. They are an important feature of HD and contribute to impairment of quality of life [6]. The most common symptoms are loss of energy and initiative, poor perseverance and quality of work, impaired judgment, poor self-care and emotional blunting. Affective symptoms such as depression, anxiety, apathy and irritability also occur frequently [7].

While there is no cure for HD, the emphasis is on the care of patients with HD, especially those in the advanced stage of the disease [2]. The aim of all treatment is on improving the quality

of life of patients with HD. Music therapy could be a complimentary, non-pharmacological intervention to reach this goal.

Music therapy

Among the many definitions of music therapy, the most used one is the one from the American Music Therapy Association (AMTA) which describes music therapy as follows:

“ Music therapy is the clinical and evidence-based use of music interventions to accomplish individualized goals within a therapeutic relationship by a credentialed professional who has completed an approved music therapy program” [8].

Music therapists assess emotional well-being, physical health, social functioning, communication abilities, and cognitive skills through musical responses. Music therapists design sessions for individuals and groups based on client needs using music improvisation, receptive music listening, song writing, lyric discussion, music and imagery, music performance, and learning through music. Finally, they are part of the multidisciplinary team and thus participate in interdisciplinary treatment planning, ongoing evaluation, and follow-up [8].

Music Therapy and Huntington’s Disease

As the ability to communicate and express oneself deteriorates over time, music therapy could play an important role in the treatment of patients with HD in all stages of the disease. Through music therapy an additional means of communication can be provided, enabling the patient to express his or her needs and emotions, and to recall life experiences which in patients with HD is also affected.

Specific literature about MT in HD is scarce. Research into the effect that music therapy has on HD began to develop in the late 1970’s and early 1980’s. Studies include: speech facilitation through singing [9], music and movement programs [10], emotional expression by use of song-writing [11], participation and communication [12]. The results of a music therapy referral audit by Daveson (2007) revealed that patients with HD were most likely to be referred to a music therapist due to a need for emotional expression, and the maintenance of communication skills and social relationships. [13]. In a comprehensive systematic literature review (see chapter 2 of this thesis), Bruggen-Rufi Van and Roos [14] conclude that music therapy interventions vary throughout the different stages of the disease and are not yet applied optimally. Not the stage of the disease, but the individual treatment goals (see table below) seem to be more determinative for the music therapy indications.

In conclusion, precise aims and methods in relation to the stage of the disease, as well as the effectiveness of music therapy, are not well determined (see table “Music Therapy and HD: Referrals and Goals” in chapter 2). There is a need for a systematic study to determine the indications to use music therapy and to study the effectiveness of music therapy in HD [14].

Despite positive findings in the field, studies linking music therapy and HD together are limited. Therefore, review of the broader field of neurodegenerative diseases that have similar symptoms to HD is essential to develop the theory building about MT and HD further.

Over the past decades, music therapy (MT) has been developed for patients with other neurodegenerative diseases, such as dementia [15,16]. There is evidence that music therapy influences emotional well-being positively and that participation in music therapy increases social response in persons with dementia, thus providing additional means of communication and enabling the patient to express his or her needs and emotions [17,18]. Through music, contact can be established, especially as language functions deteriorates during the later stages of the dementia process [19]. Furthermore, enhancing the ability for self-expression, contributing to improvement of the quality of life, has been reported by Lee and McFerran who describe five females in a multiple case study with profound and multiple disabilities using patient preferred song-choices in music therapy [20]. The patient can be stimulated to recall life experiences through music. In the music therapy session this can be used as a catharsis to experience emotions. In a recently update Cochrane review on music-based therapeutic interventions for people with dementia [21] the authors conclude that music therapy may reduce depressive symptoms.

Based on the above-mentioned benefits, the assumption can be made that MT is beneficial to patients with HD on communicative and expressive skills. Improvement of these skills might lead to improvement of behavior, eventually leading to improvement of the quality of life of patients with Huntington’s disease [14].

Aims and outline of this thesis

The specific aim of this thesis was to explore the effectiveness of music therapy in patients with HD. To achieve this goal, the first step was to study the available literature on music therapy in HD. The result of this literature search is described in **chapter 2**.

Subsequently, we conducted a focus group study to investigate whether six professionals from different disciplines, who all have a considerable amount of years of clinical experience with patients with HD, recognized a possible beneficial role for the music therapist in relation to improvement of the quality of life for people with HD. The outcome of this study is described in **chapter 3**.

Both the literature and the focus group study helped us to design a randomized controlled trial to study the effect of music therapy in HD. The protocol of this RCT is described in **chapter 4**. The study and the results of the RCT are described in **chapter 5**.

Performing a multi-center RCT studying the efficacy of music therapy with vulnerable patients in four different long-term care facilities is considered to be a complex intervention. In order to elucidate the results and to investigate how the study was performed, we conducted a process-evaluation. This process-evaluation, described in **chapter 6**, highlighted the many barriers and facilitators to perform such a complex intervention, and the outcome helped us to elucidate the results of this quantitative effectiveness study, resulting in recommendations for future studies in regard to design and measurement tools.

In **chapter 7**, two case reports with two patients who benefit from music therapy are described for illustration. At the same time, these case examples give the reader some insight in the real-world context in which music therapy is practiced.

Finally, the main conclusions of this thesis are summarized and discussed in **chapter 8**. In this chapter we will build on the definitions of music therapy to determine whether the methods that we used throughout this thesis were in compliance with these descriptions, followed by future perspectives and closing remarks.

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